PEDIATRIC TUMEFACTIVE MULTIPLE SCLEROSIS – A CHALLENGING CASE

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ABSTRACT

Multiple sclerosis (MS) is an autoimmune inflammatory disease of the central nervous system which is often diagnosed in young adults. We present a case of tumefactive multiple sclerosis in a child, an unusual form of presentation of MS, especially at this age group, delineating the clinical picture, the diagnostic difficulties and the possible therapeutic options we have gathered.

Keywords: multiple sclerosis, child, tumefactive

INTRODUCTION

Multiple sclerosis (MS) is an autoimmune inflammatory demyelinating disease of the central nervous system. It is tipicallyencontered in young adults and considered a rare occurence in children, especially before the age of 10. There are, however, increasing cases described among children, despite the difficulty in uniting the diagnostic criteria.

Clinically, MS is characterized by recurrent episodes of neurological impairment, separated in time and space. It should be noted that young children can initially show multifocal neurological deficits and encephalopathy, making a differentiation from ADEM (acute disseminated encephalomyelitis) difficult.

According to the 2010 Revised Diagnostic Criteria for MS (1), which have included pediatric patients into the report, the diagnosis of MS requires one of the following conditions: two or more nonencephalopathic central nervous system events with presumed inflammatory cause, separated by more than 30 days and involving more than one area; or one nonencephalopathic episode typical of MS which is associated with MRI findings consistent with 2010 McDonald criteria for dissemination in space and in which a follow-up MRI shows at least one new enhancing or nonenhancing lesion consistent with the criteria for dissemination in time; or one ADEM attack followed by a nonencephalopathic event, three or more months after symptom onset, that is associated with new MRI lesions that fulfill the 2010 McDonald dissemination in space criteria; or a first single acute event that does not meet ADEM criteria and whose MRI findings are consistent with the 2010 McDonald criteria for dissemination in space and dissemination in time (applies only to children over 12 years old).

Dissemination in space (1) consists in one or more T2 lesions in at least two of four MS-typical regions of the central nervous system: periventricular, juxtacortical, infratentorial or spinal cord.

Dissemination in time (1) consists in the simultaneous presence of asymptomatic gadolinium-enhancing and nonenhancing lesions at any time, or a new T2 and/or gadolinium-enhancing lesion on

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follow-up MRI, irrespective of its timing with reference to a baseline scan.

The cerebrospinal fluid analysis in patients with MS shows oligoclonal bands and/or an increased IgG index.

Tumefactive multiple sclerosis represents an unusual presentation, a tumor – like variant of MS (2,3), with large lesions often associated with edema. While it represents a rare diagnosis in adults, it is even more uncommon in children.

We describe a case of a progressive demyelinating disease in a child whose clinical and imagistic presentations, together with the CSF analysis were ultimately united into the diagnosis of tumefactive multiple sclerosis.

CASE REPORT

On awakening, in the morning, a ten year old male patient, T.S., known with no significant personal or family history, suffered a sudden left hemiparesis, associated with a left central facial palsy.

He was, at that time, investigated in a neurosurgery department. The cerebral MRI revealed one right frontal paraventricular lesion with a diameter of two cm, displaying only mild contrast enhancement, with no associated perilesional edema, which was responsible for the child's clinical signs (Fig. 1).

However, the MRI also showed multiple infra- and supratentorial white matter lesions which were at first thought to be of demyelinating nature (Fig. 2).

Consequently, a stereotactic brain biopsy was performed at the site of the right frontal lesion described above. The histopathological examination carried out in the same clinic pleaded for a demyelinating lesion. On further analysis, the histological assessment using immunohistochemistry identified the lesion as being a ganglioglioma and prompted the child towards a neurosurgical excision procedure. The histopathological analysis of the tissue extracted from the lesion was again suggestive of ganglioglioma.

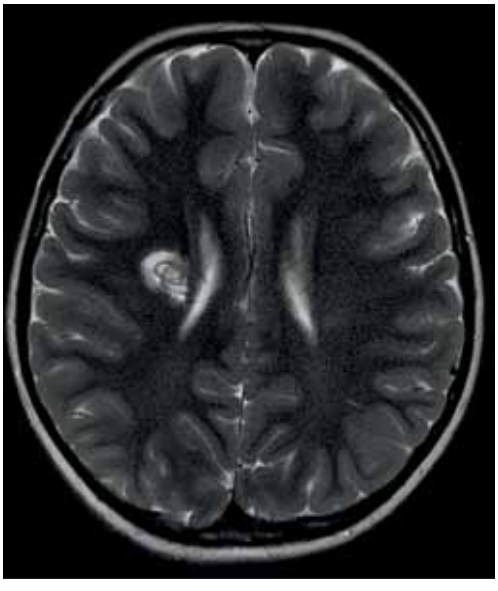

FIGURE 1. *T2 – pondered Axial Cerebral MRI, right frontal periventricular white matter expansive lesion.*

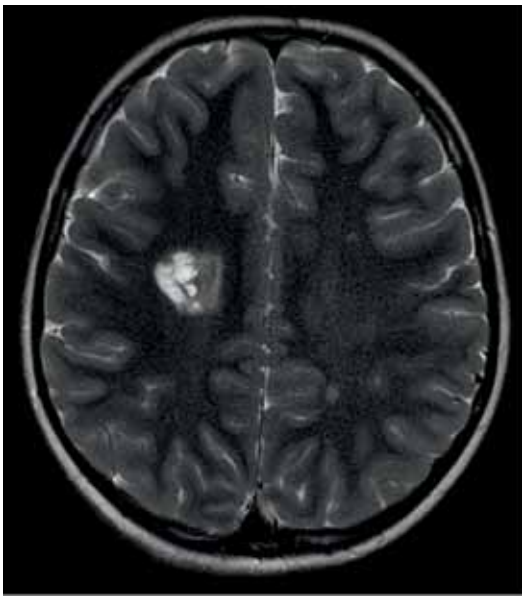

FIGURE 2. *T2 – pondered Axial Cerebral MRI, multiple left fronto-parietal subcortical and deep white matter lesions.*

On admission to our clinic, the child's neurological examination revealed postoperatorysequelar left spastic hemiparesis and mild left central facial palsy. He had suffered no other neurological deficit since the onset of the disease.

However, seriated cerebral imagistic examinations revealed a numerical expansion of the cerebral lesions, one of them being spotted at C1 medullary level. This oriented the diagnosis towards a demyelinating disease, despite the controversial results of the repeated histopathological examinations.

Further on, a lumbar puncture was performed, showing the presence of oligoclonal bands in the cerebrospinal fluid, which also supported the diagnosis of MS.

Consequently, a follow-up imagistic examination was performed, showing the appearance of small new lesions at the C2 medullary level, as well as in the brainstem (Fig. 3 and Fig. 4).

Given the imagistic signs of progression of the disease, the patient was started on corticotherapy, for one month, with no new clinical events. Cur-

FIGURE 3. *T2 pondered Sagital Cerebral and Spine MRI – multiple white matter lesions involving the brainstem and the upper cervical spine.*

FIGURE 4. *Close-up of cervical lesion*

rently, the child is beingclinically monitored on a three month period and we plan on obtaining an MRI every six months. Also, he is undergoing periodical ophtalmological evaluations, including measurement of visually evoked potentials, which up to this point were normal.

Corticotherapy is the first line treatment choice in case of a new acute clinical event (4). Other therapeutic options include intravenous immune globulins. There is little experience in children with the long term immunomodulator agents such as interferon and glatiramer (5), these being treatment options to be considered with time, according to the patient's clinical and imagistic evolution.

CONCLUSIONS

We have presented a case of acute motor deficit, with an unusual imagistic aspect, which was finally diagnosed with tumefactive multiple sclerosis, a rare diagnosis, especially in children. Despite being recognized as a disease mainly affecting young adults, multiple sclerosis should also be taken into consideration when evaluating children. A thorough imagistic investigation needs to be carried out in order to assess the presence of demyelinating lesions. CSF analysis can be of aid in the diagnosis, if it shows the presence of oligoclonal bands or a high IgG index. The patients need to be on a long term follow-up with a multidisciplinary team consisting in pediatric neurologists, ophtalmologists, radiologists and pediatricians.

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